



Case Presentation

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Clinical Background



- 39 year old smoker lady, presented with a 6 month history of toothache and headache
- Medical history: hypertension

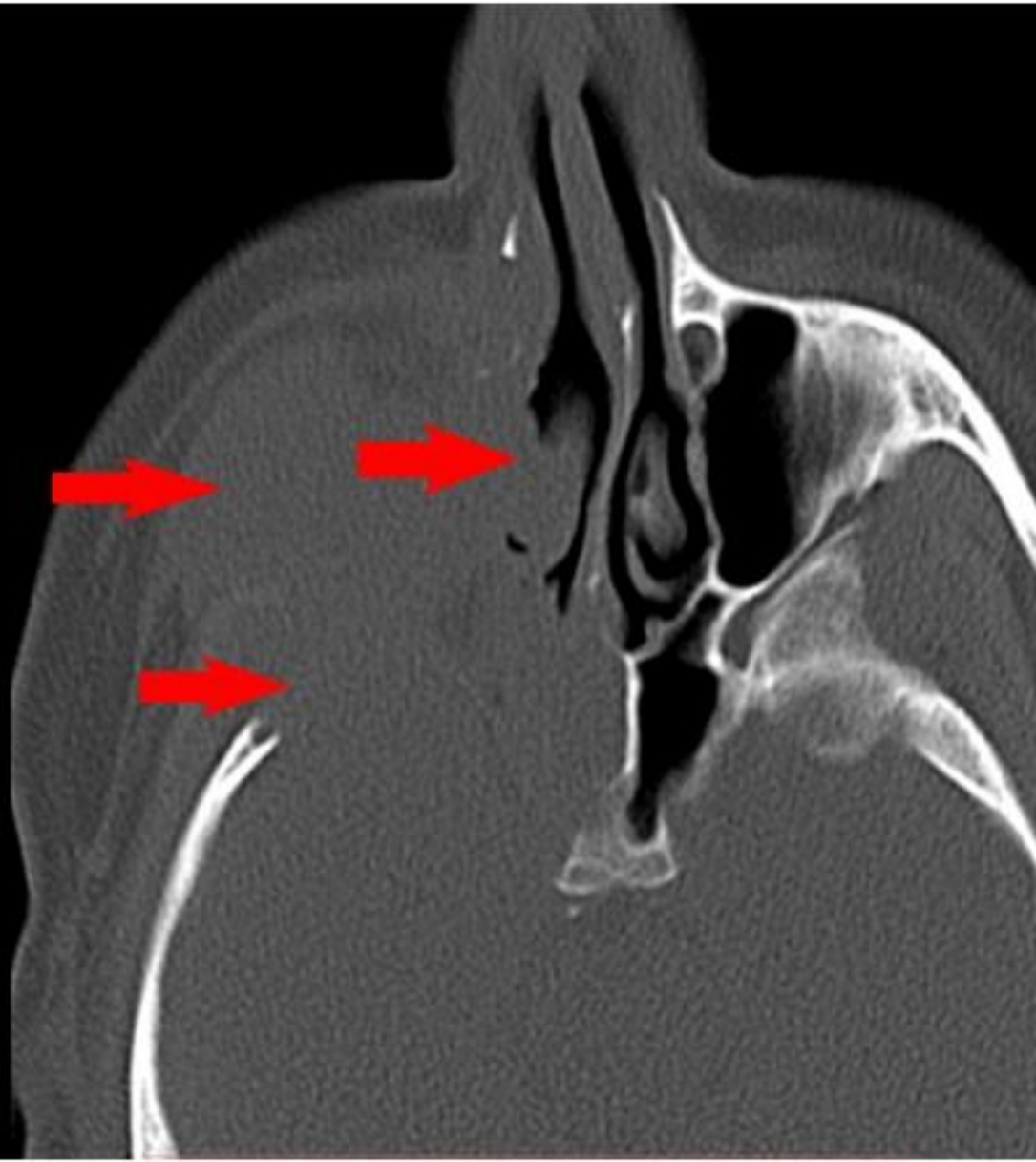


Clinical Background



- Multiple visits to dental clinics, received pain killers but without relief
- Visited an ENT clinic, physical exam revealed a nasal cavity mass
- CT scan was performed

CT scan

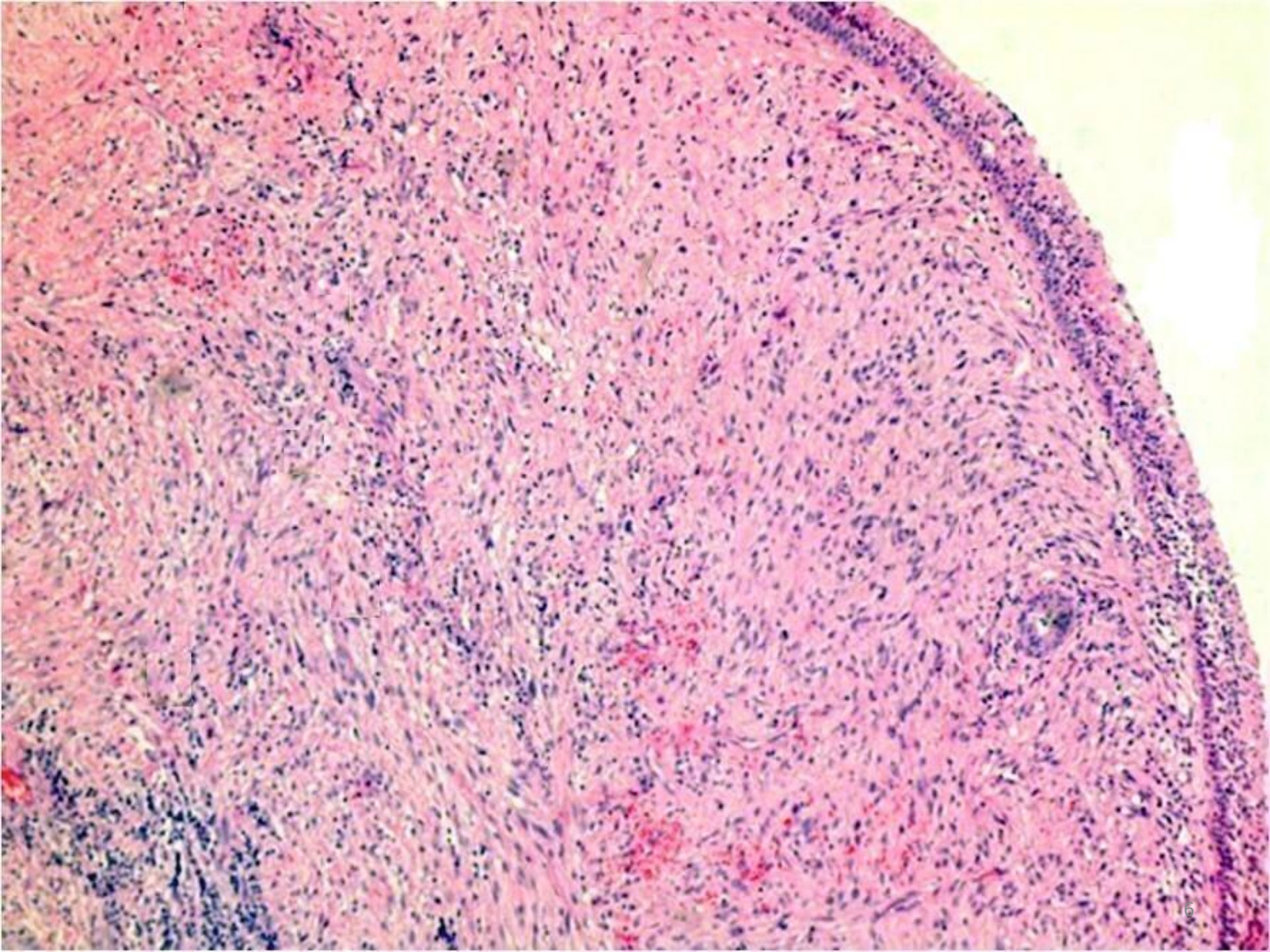


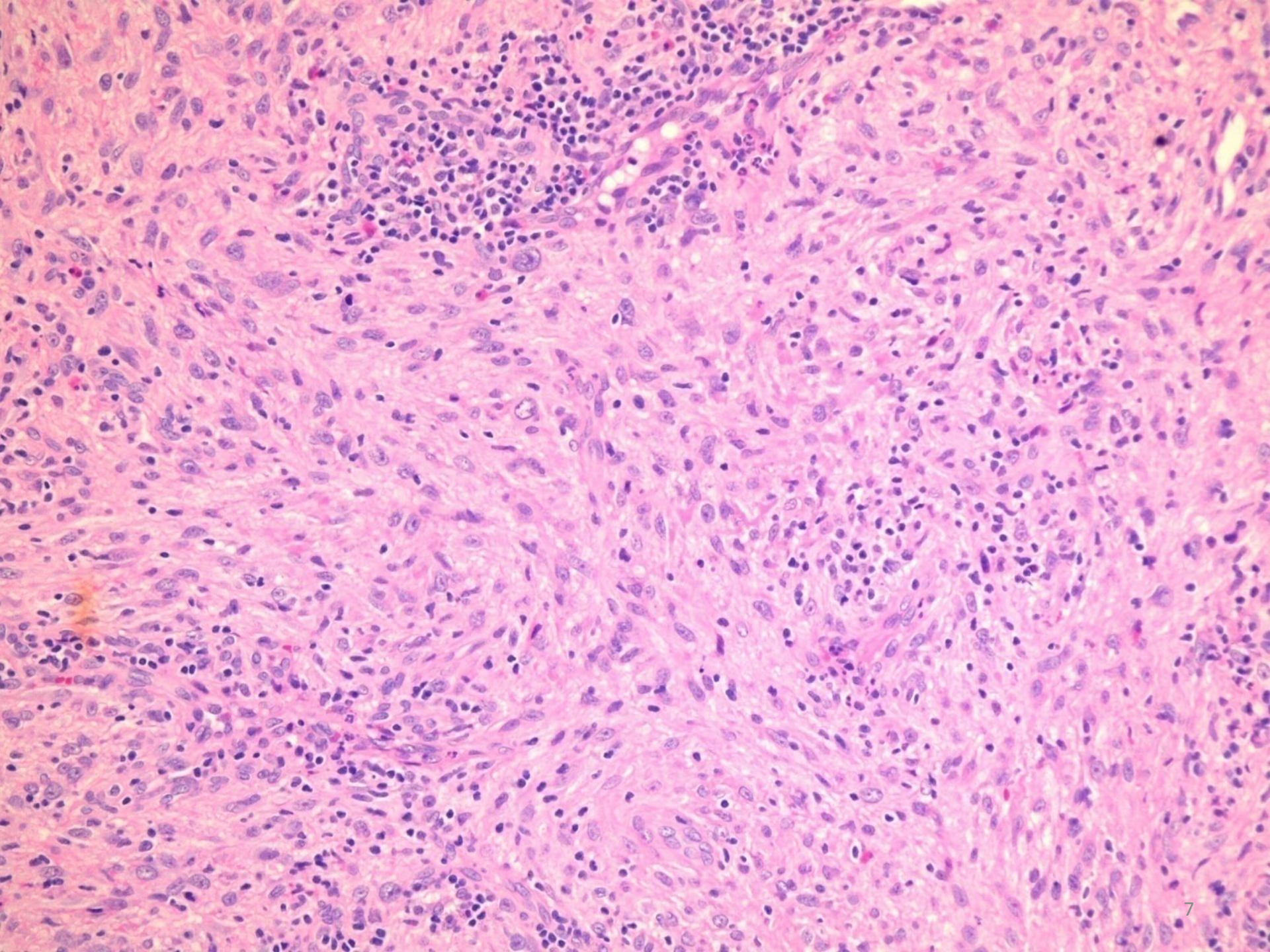


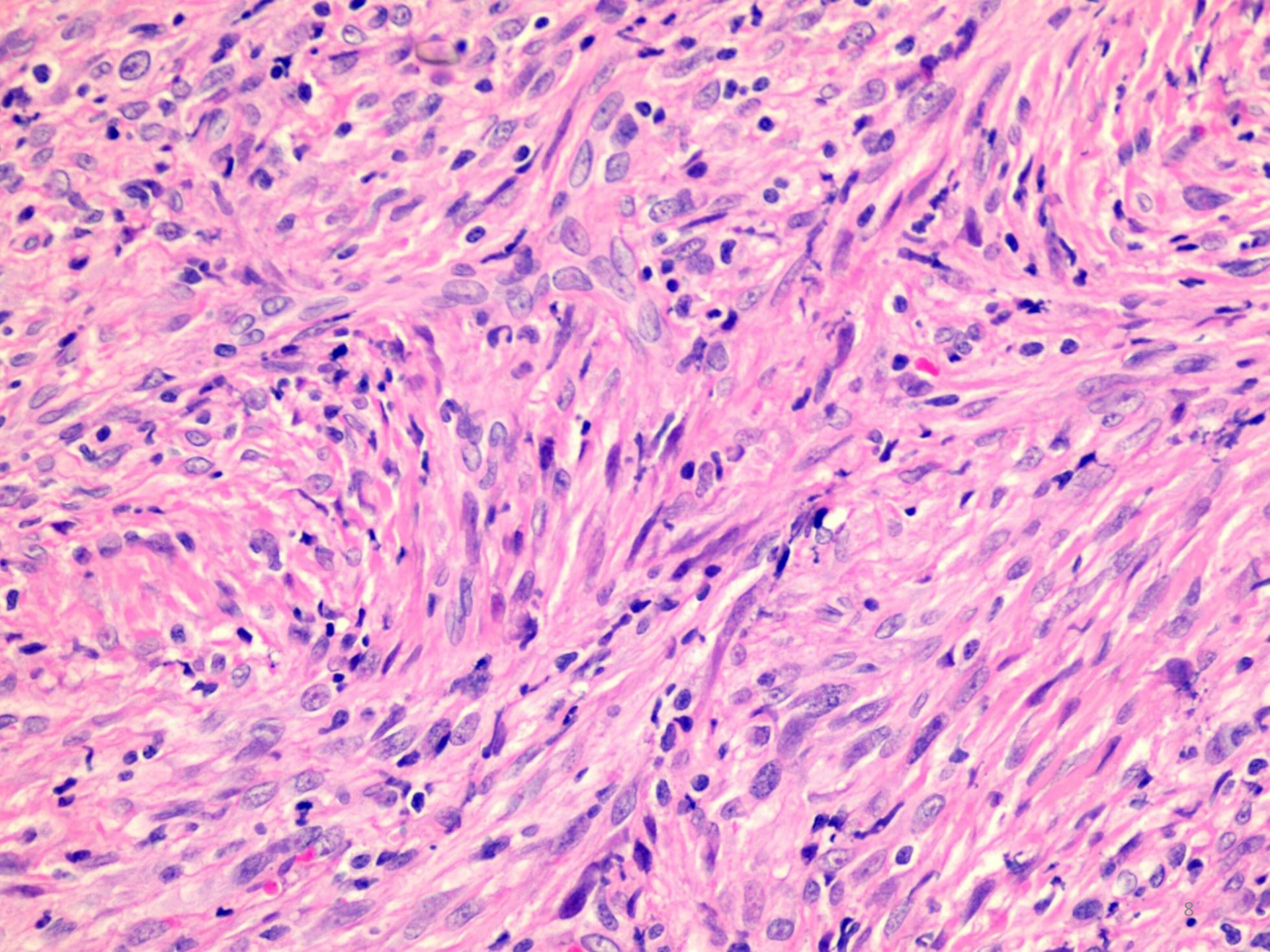
Radiologic findings

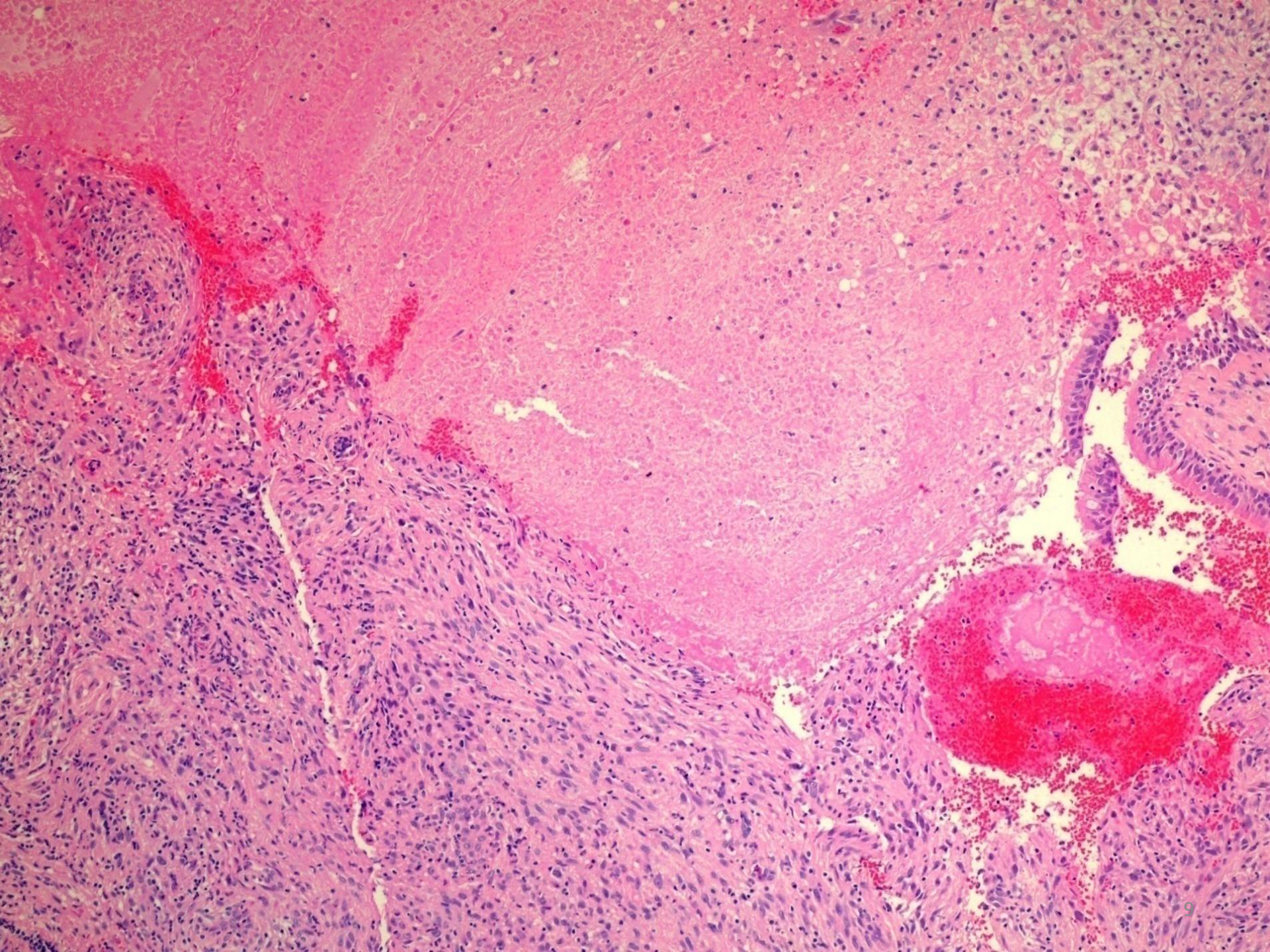


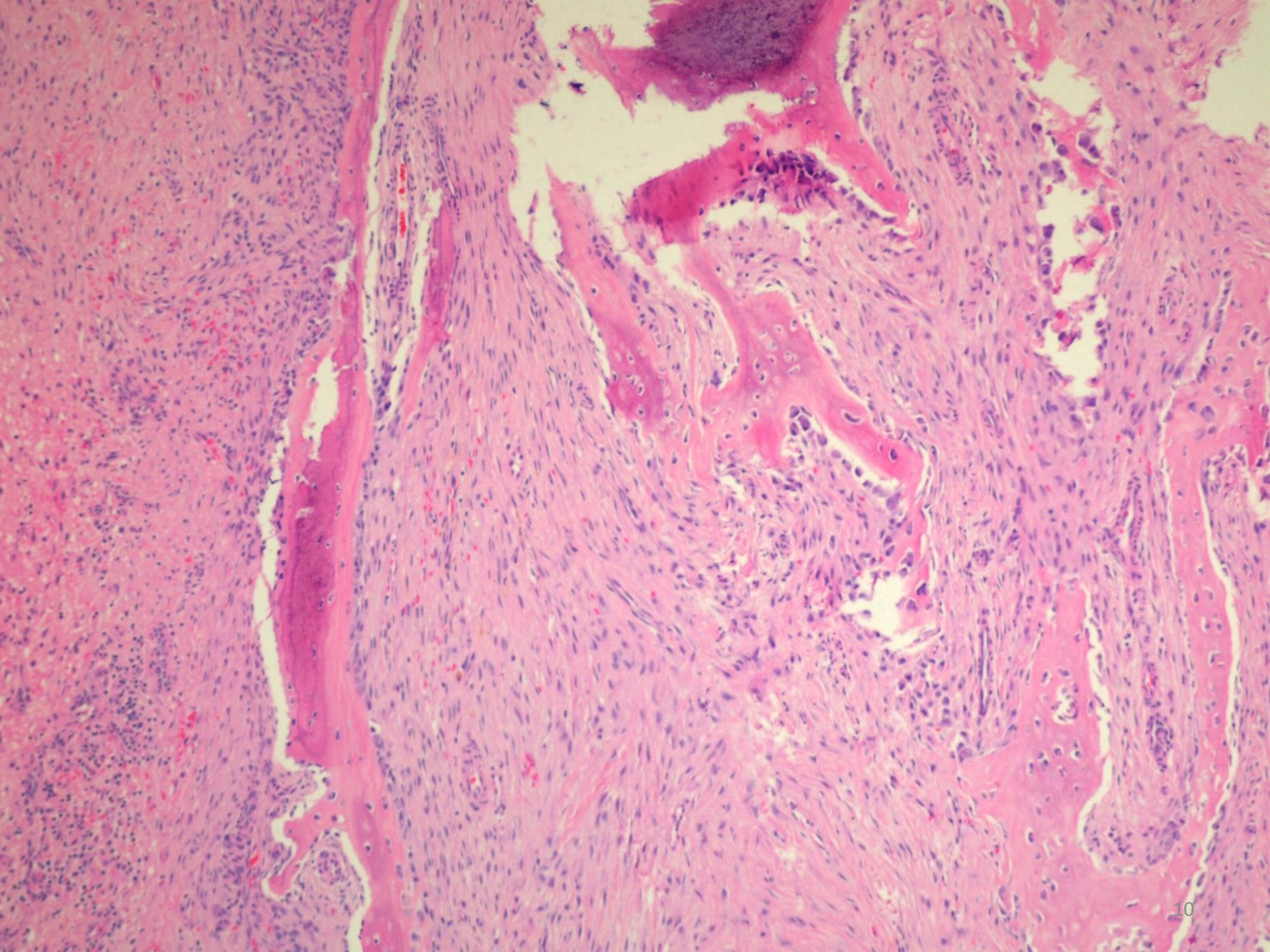
- Large, lobulated, destructive solid tumor measuring 5 cm in maximum dimension, involving the roof of the mouth, the right nasal cavity, maxillary sinus, orbit
- The tumor also extends to the right infratemporal fossa, pterygopalatine fossa and parapharyngeal space, sparing the oropharynx
- The patient underwent endoscopic biopsy from the nasal mass

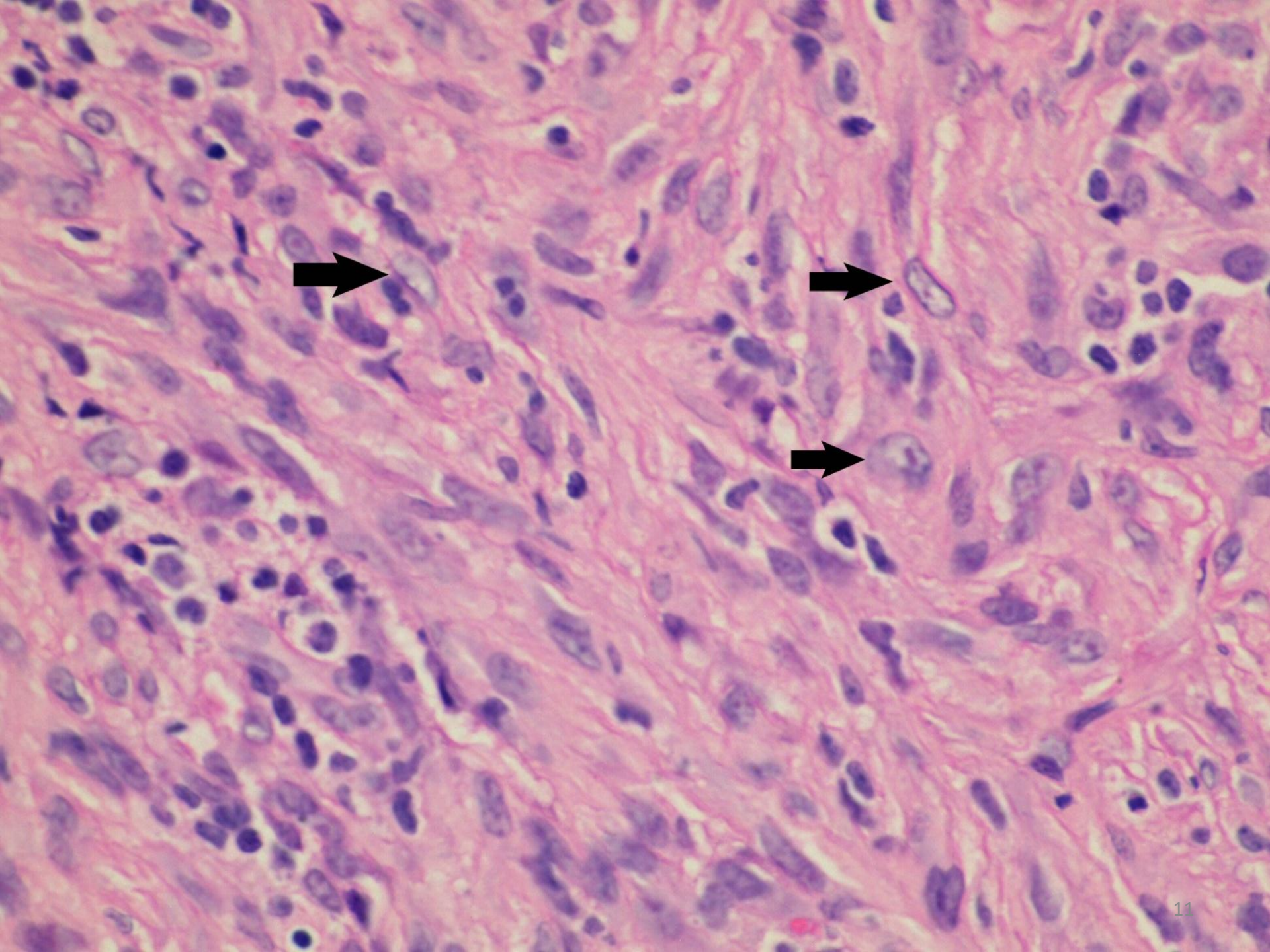


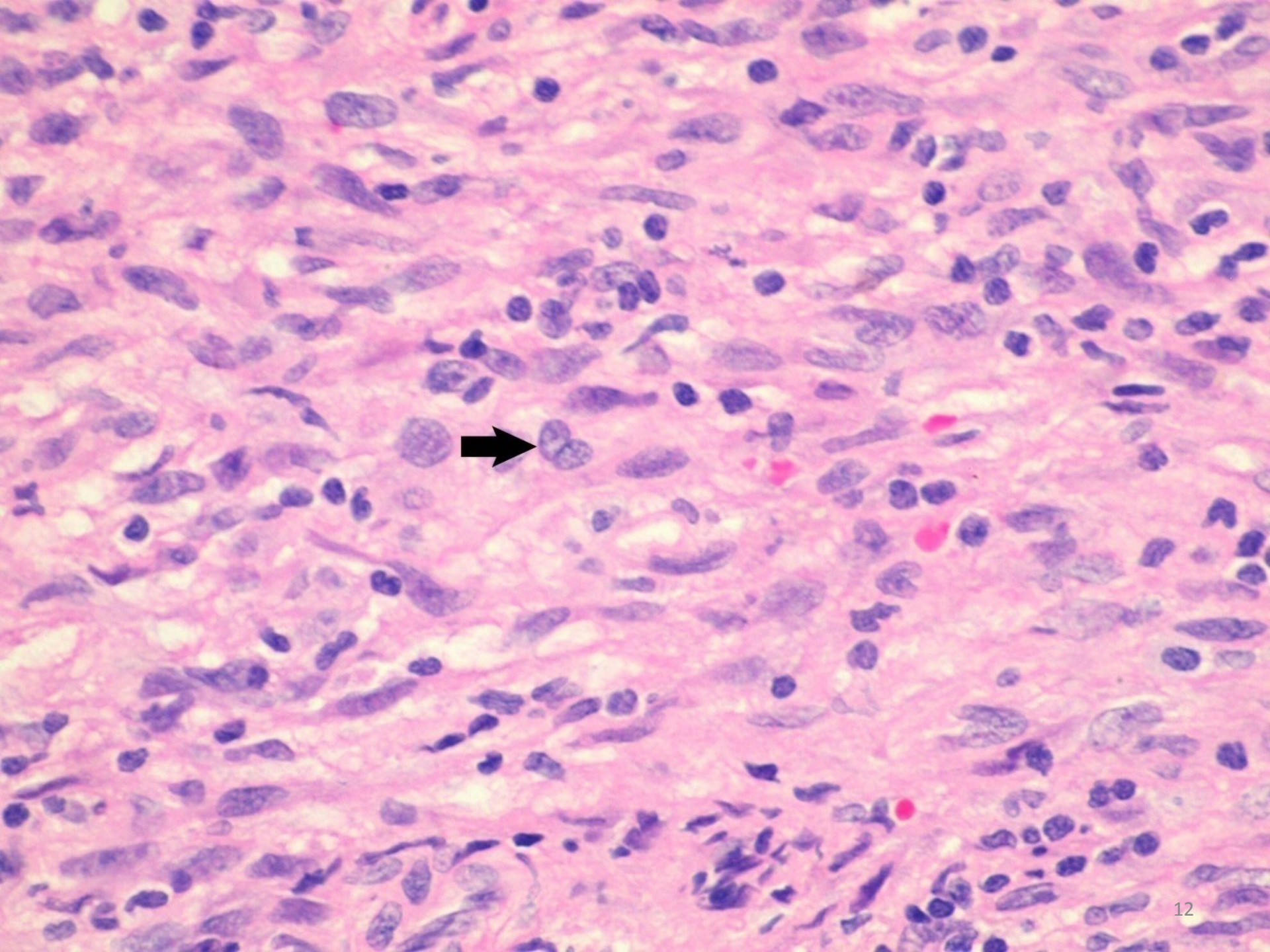












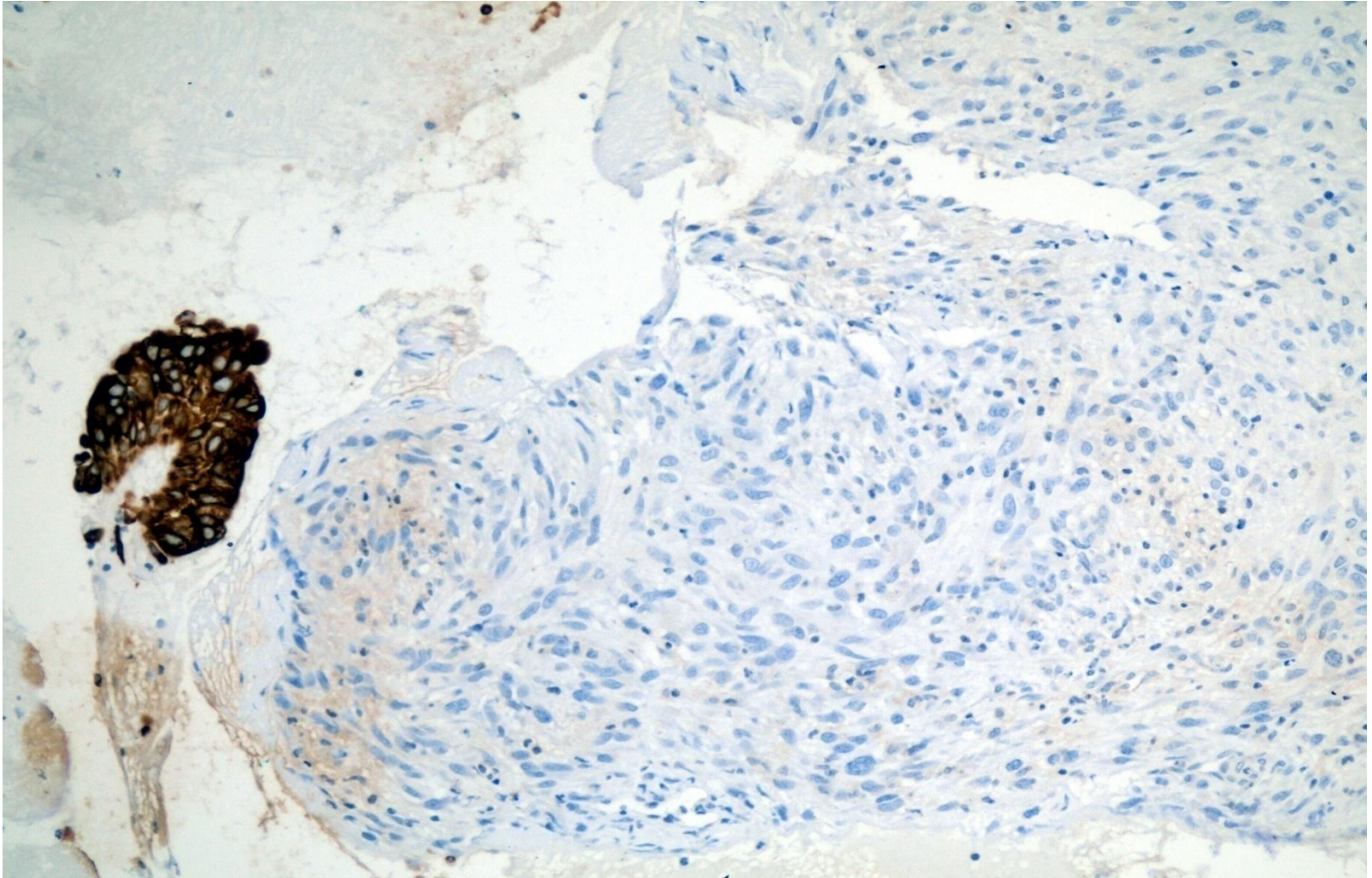


Differential Diagnosis

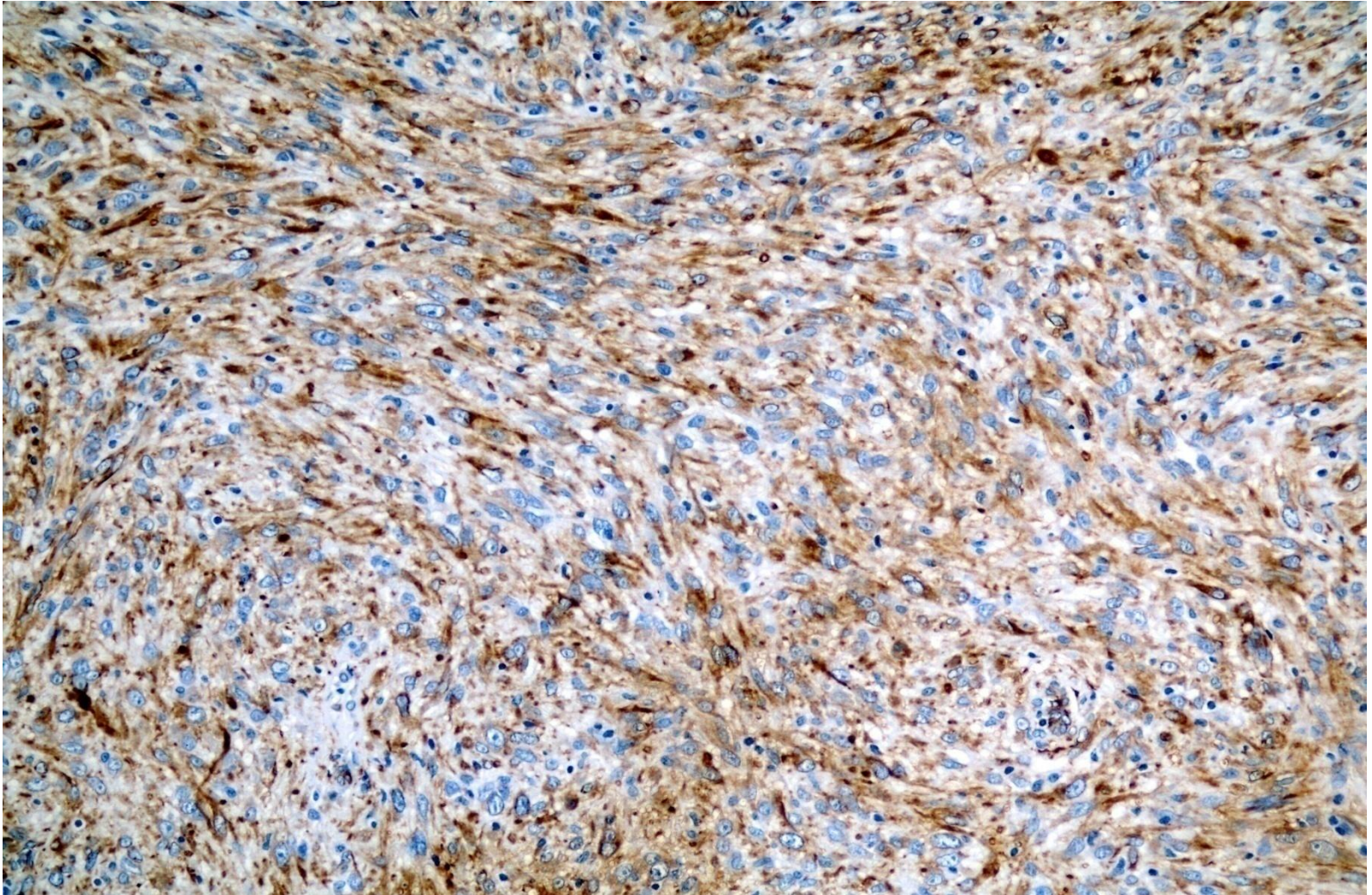


- Sarcoamntoid carcinoma(CK??)
- Extracranial meningioma (EMA, vimentin??)
- Leiomyosarcoma (Desmin, SMA??)
- Solitary fibrous tumor (Bcl2, EMA??)
- Anaplastic large cell lymphoma(CD30,CD45??)
- Melanoma (HMB45??)

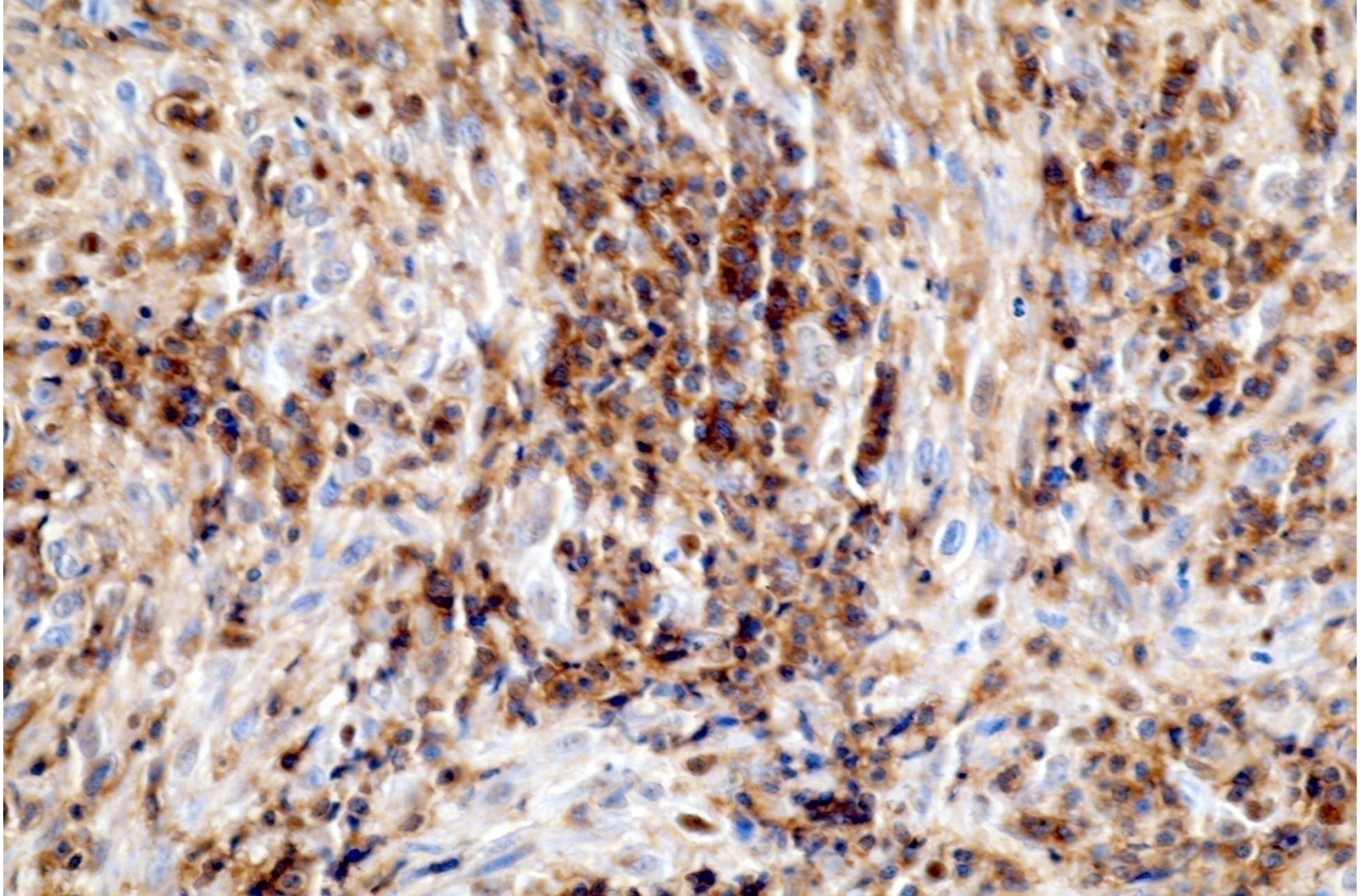
cytokeratin



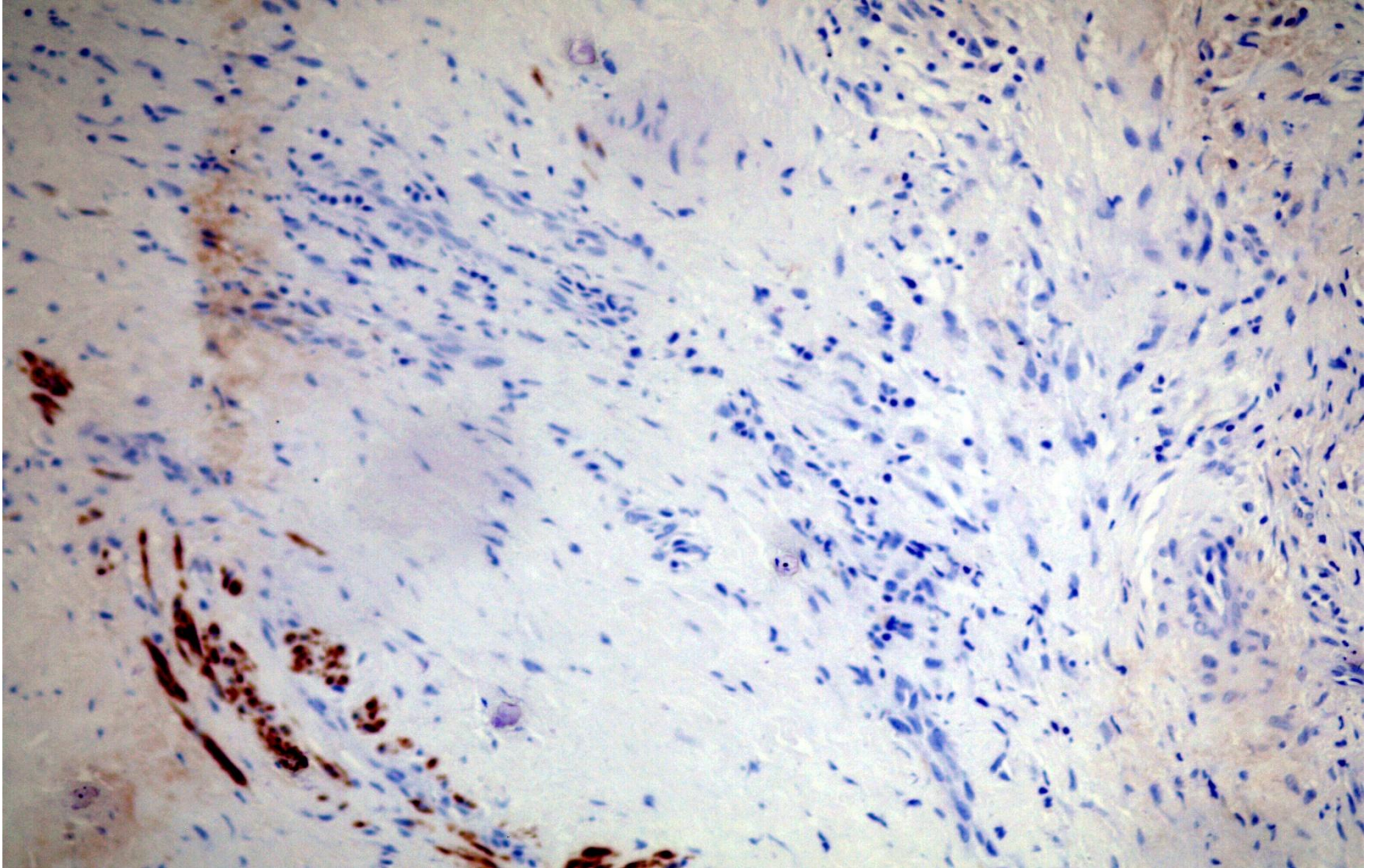
vimentin



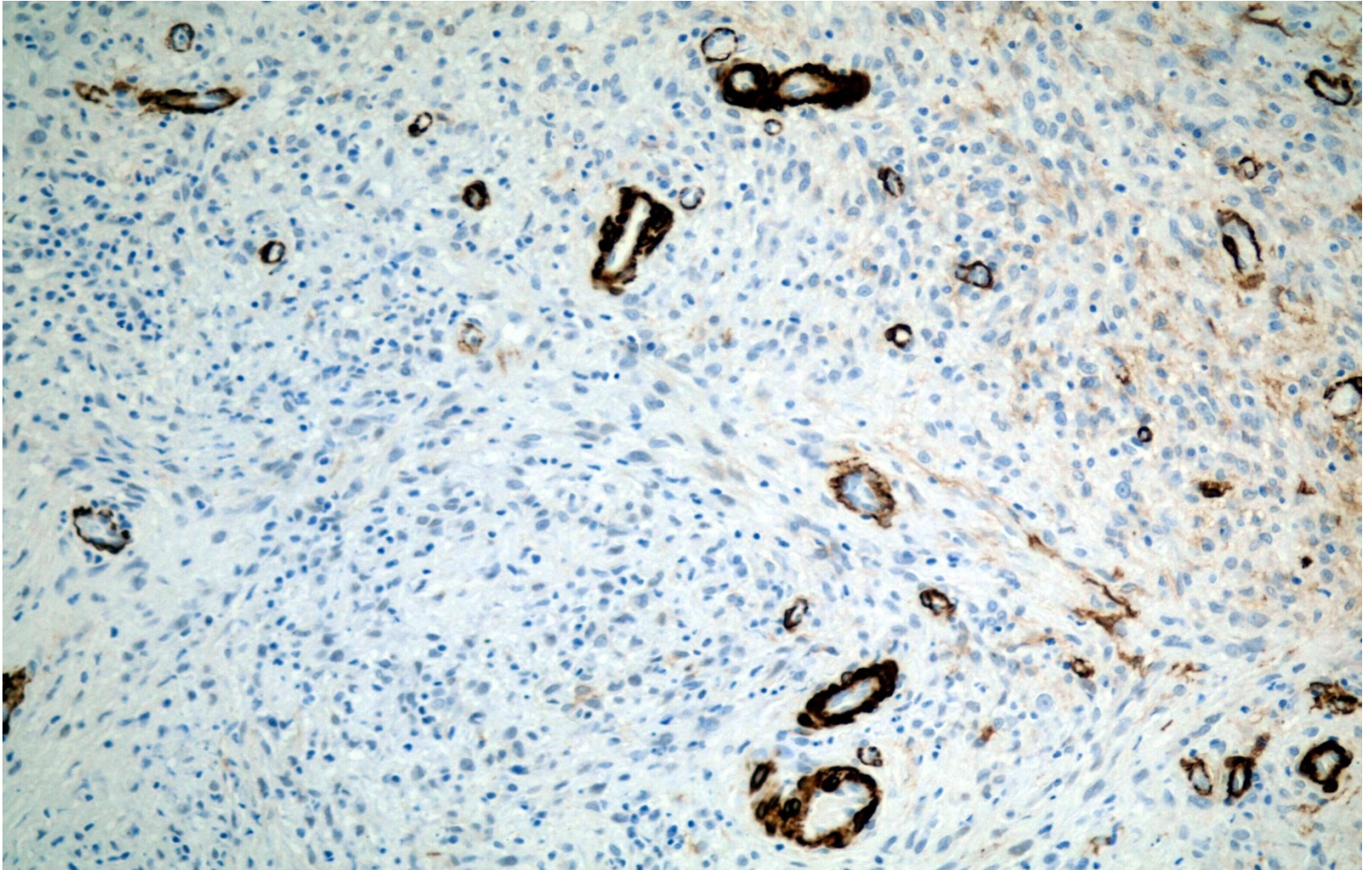
CD45



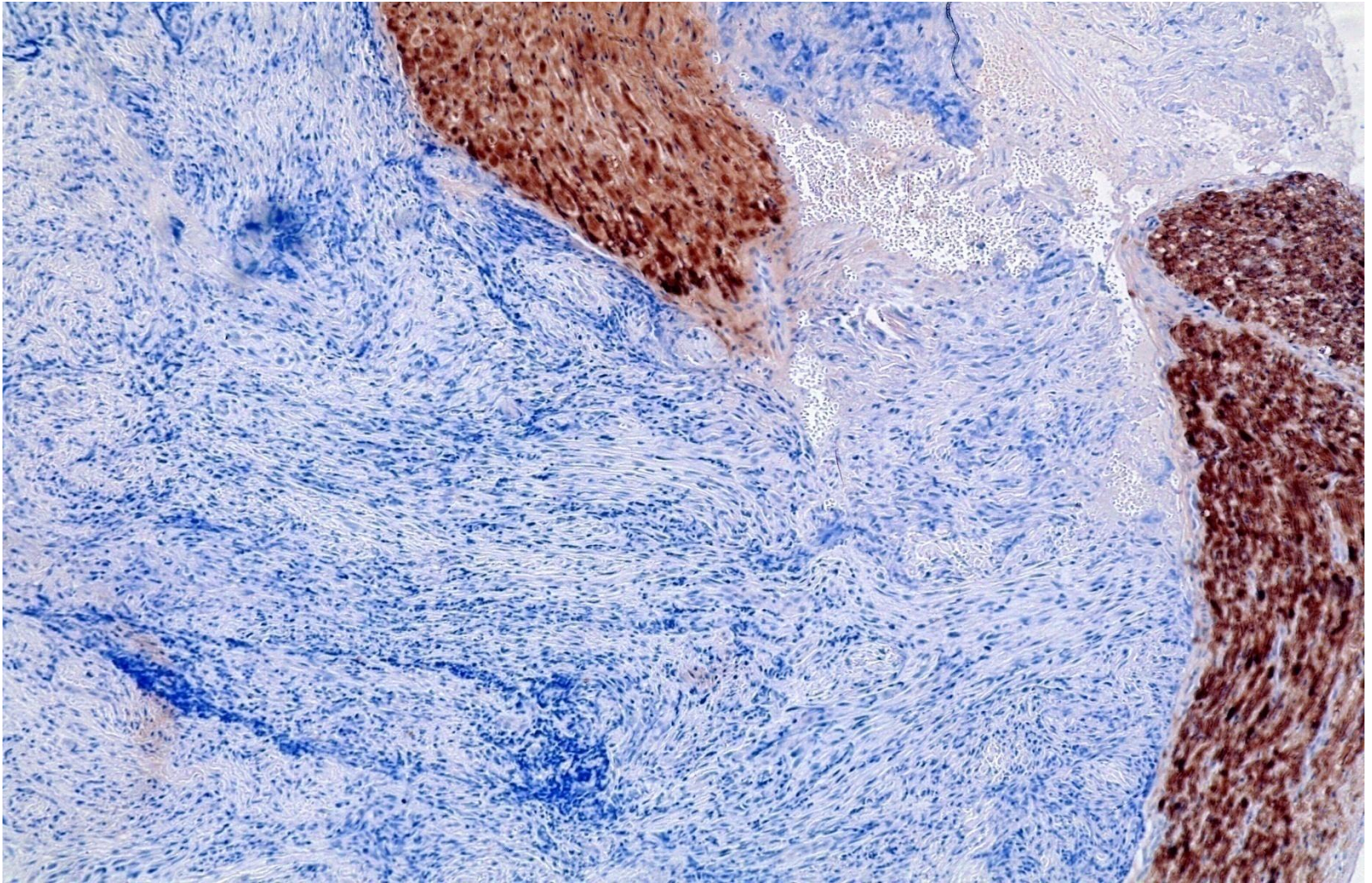
SMA



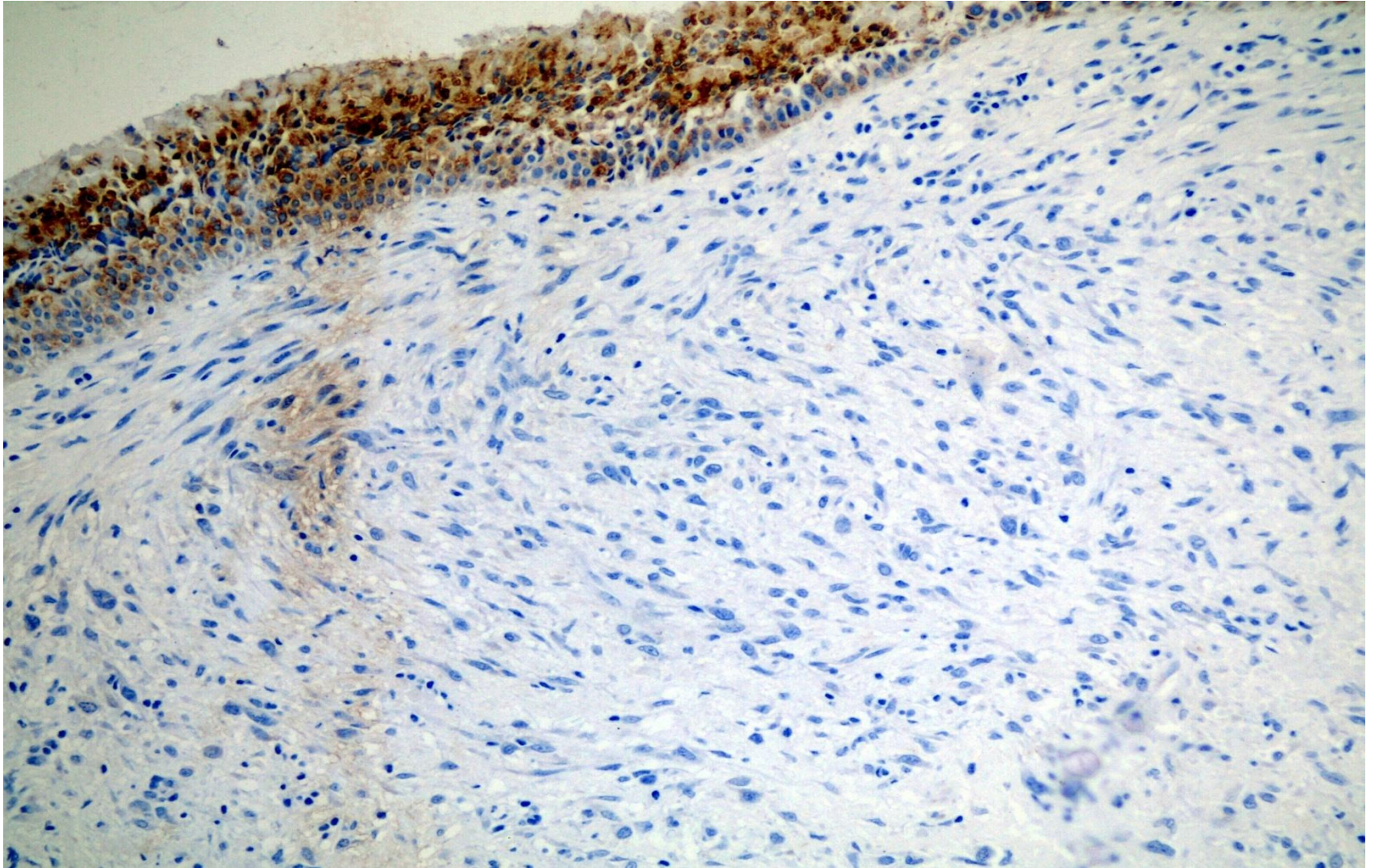
Desmin



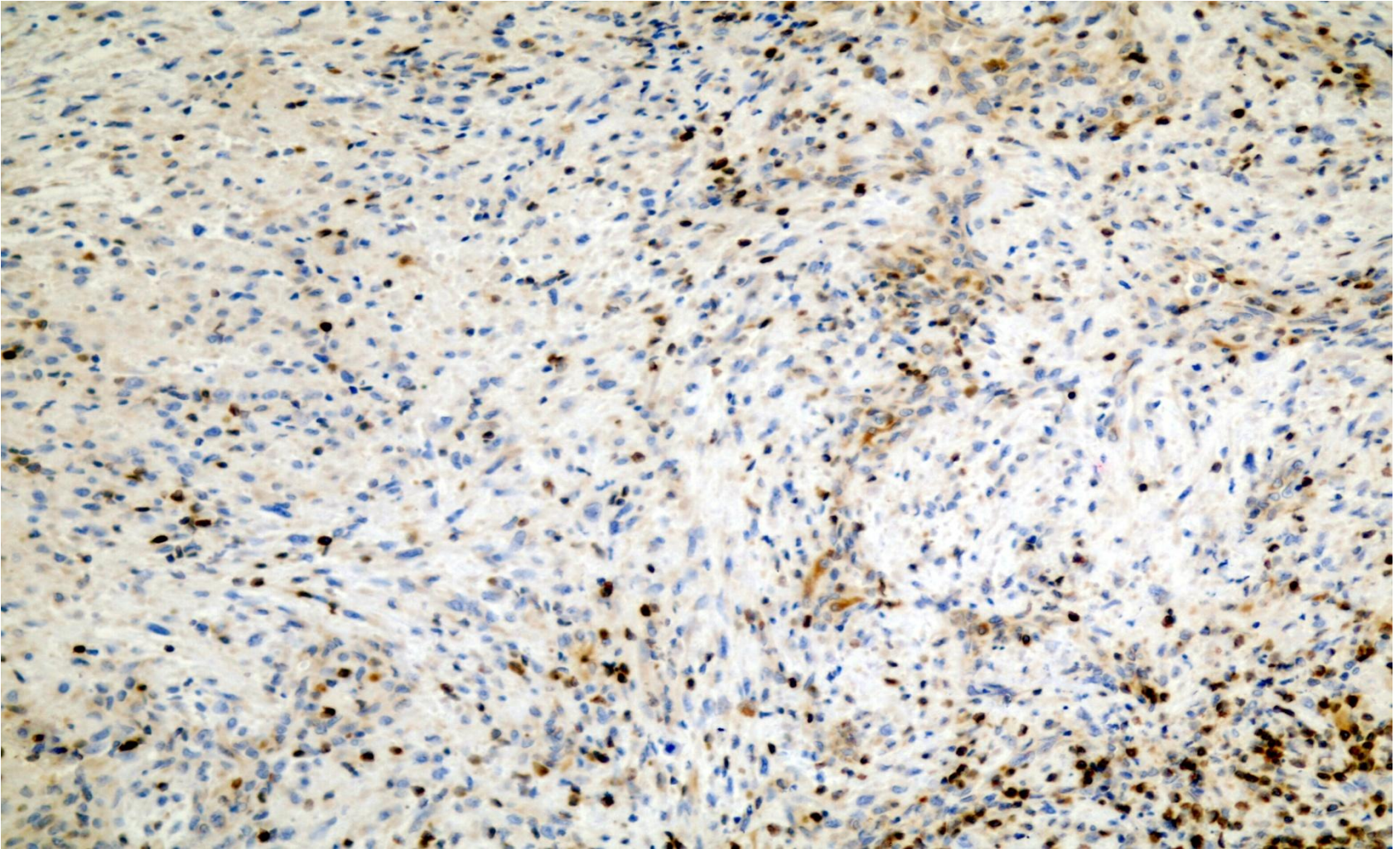
S100



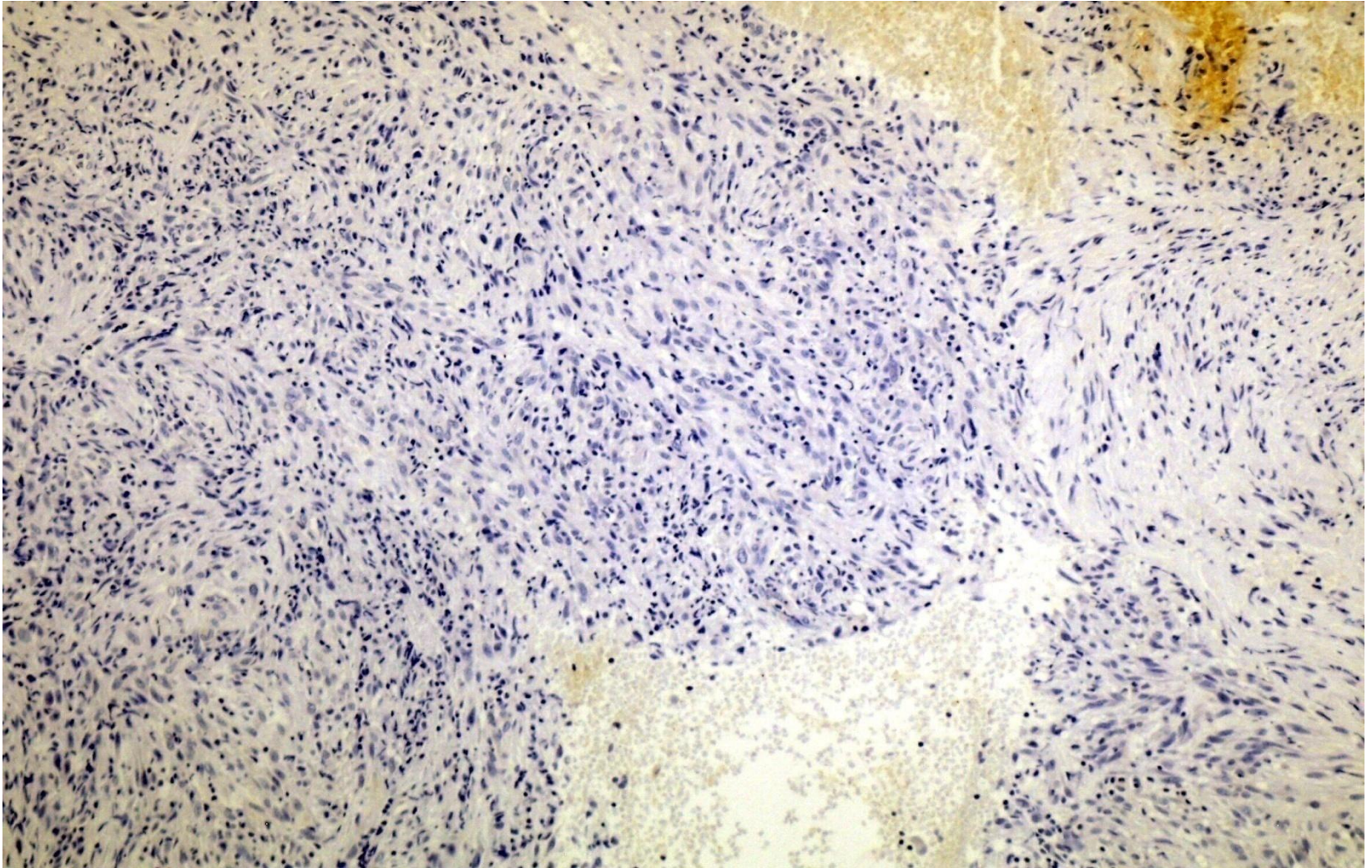
EMA



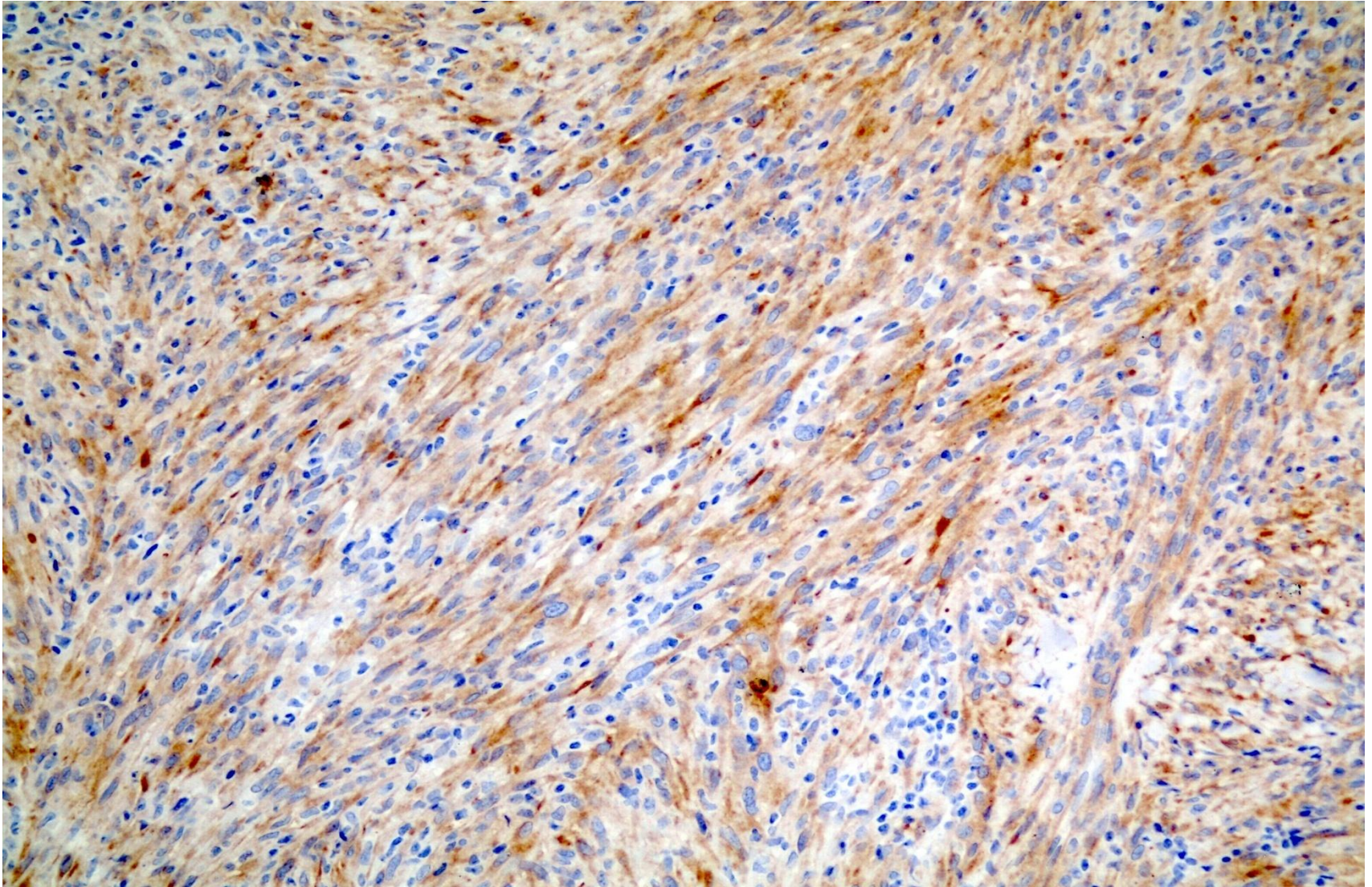
Bcl2



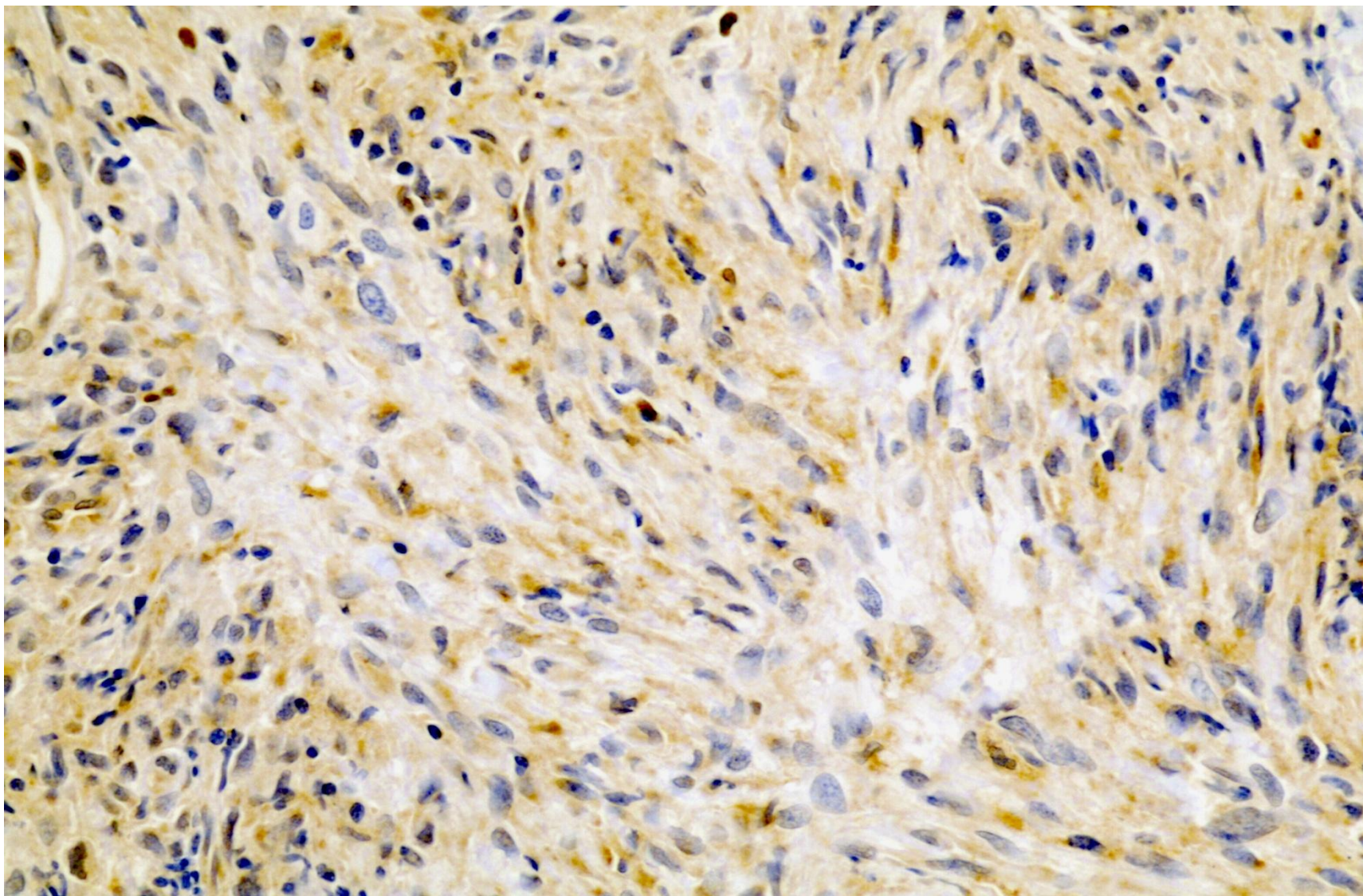
CD30



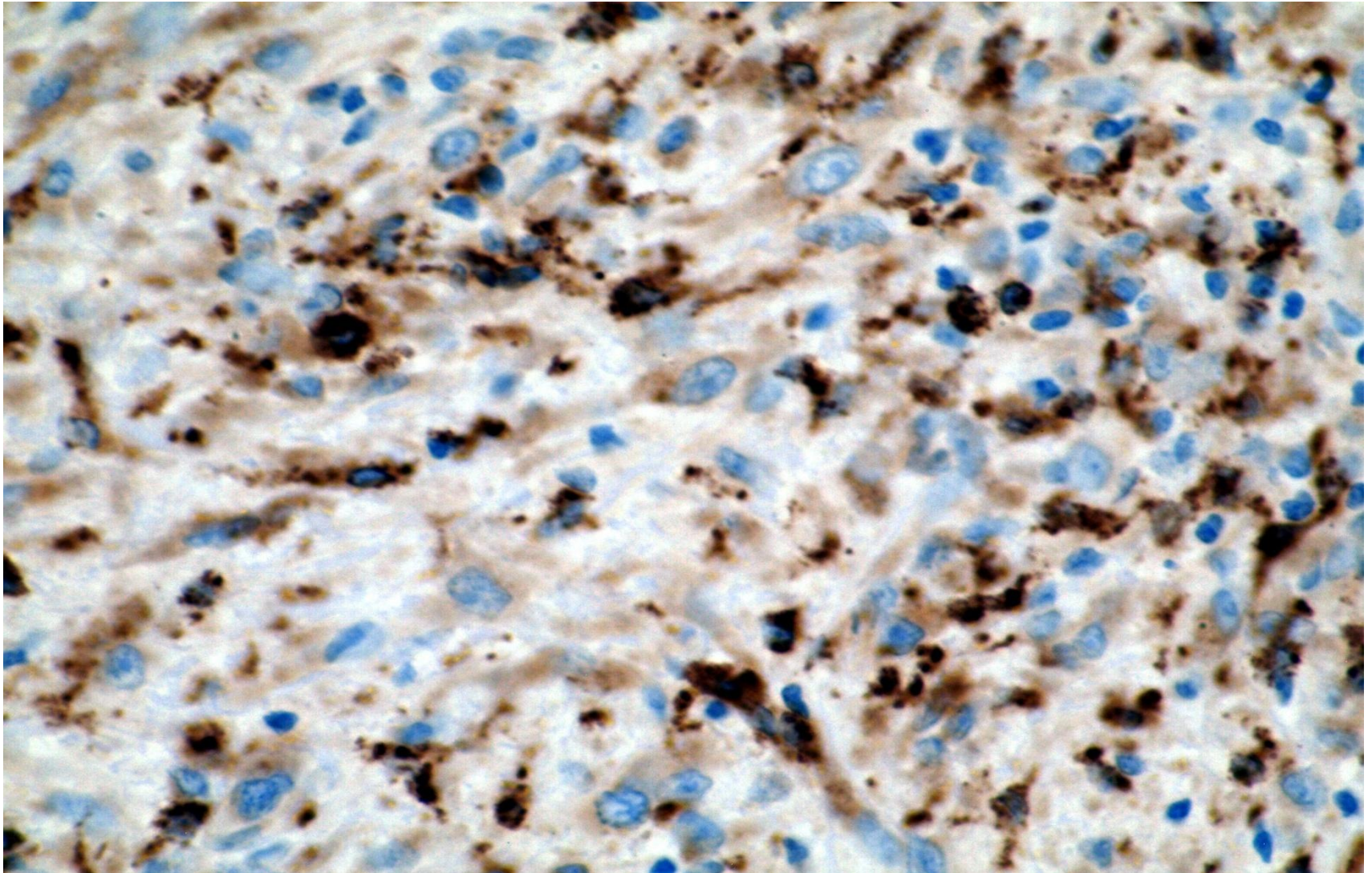
CD21



CD23



CD68





Immunoprofile



Positive

- vimentin
- CD21
- CD23
- CD68 (focal)

Negative

- S100
- CD45
- CK
- EMA
- SMA
- Desmin
- HMB45
- CD99
- Bcl2
- CD20
- CD30
- ALK
- CD1a



Diagnosis

Extranodal follicular dendritic cell sarcoma



FDC sarcoma



- A rare neoplastic proliferation of FDCs (antigen-presenting immune accessory cells that are widely distributed in tissues)
- First reported by Monda et al, in 1986
- Almost all patients are adults, with a median age of 40 years and with no sex predilection
- Some association with Castleman disease (hyaline vascular variant)



Etiology



- Most cases: unknown
- Hyaline-vascular Castleman disease (10-20%) of cases
- A pathway for tumor evolution: FDC hyperplasia, dysplasia then transformation
- Overexpression of p53 protein is noted in FDC sarcoma, and is weakly positive in spindle cells in Castleman disease
- Epidermal growth factor receptor expression



Clinical symptoms



- Patients often present with a slow-growing, painless mass
- Constitutional symptoms and paraneoplastic syndromes are rare, unless associated with Castleman disease, or in inflammatory pseudotumor variant of FDC sarcoma
- Abdominal tumors present with pain and compression symptoms



Nodal vs Extranodal sites



- Nodal FDC sarcoma accounts for more than two thirds of cases, with cervical lymph nodes being the most commonly affected
- Extranodal FDC sarcoma is very rare (37 cases by 2010)*
- Most of the prototype: oropharynx
- IPT-variant: spleen or liver
- Other reported sites: GIT, soft tissue, skin, lung and breast

* Duan GJ, Wu F, Zhu J, Guo DY, Zhang R, Shen LL, Wang SH, Li Q, Xiao HL, Mou JH, Yan XC, Extranodal follicular dendritic cell sarcoma of the pharyngeal region: a potential diagnostic pitfall, with literature review, Am j clin pathol, 2010 jan.



Gross features



- Size range from 1 to 20 cm maximum diameter, with a median size of 6 cm
- Round to ovoid, well-circumscribed, fleshy masses that on cut section are solid and tan
- Interspersed areas of hemorrhage and yellowish necrotic areas might be present particularly in larger tumors



Microscopic features



- The tumor is characterized by a proliferation of spindle to ovoid cells that form fascicles, storiform patterns and whorls (360°)
- Cells tend to be plump, with slightly eosinophilic, fibrillary cytoplasm and indistinct cell borders
- The nuclei are elongated, unevenly spaced, small distinct nucleoli
- Occasional nuclear pseudo-inclusions



Microscopic features



- Small lymphocytes are scattered throughout the tumor and perivascular cuffing
- Occasional binucleated and multinucleated tumor cells may be present
- Although highly cellular, atypia is only mild to moderate
- Mitotic figures: 0-10/ 10HPF
- Necrosis can be seen in higher grade tumors

IPT variant



- Spleen or liver
- Prominent lymphoplasmacytic infiltrate, masking neoplastic cells
- Atypia is more variable within the tumor
- Necrosis and hemorrhage are more common
- EBV positive (EBER-ISH)
- More aggressive



Immunophenotype



- Normal FDC antigens: CD21, CD35, CD23, D2-40, CXCL-13
- Common positivity: vimentin, fascin, desmoplakin, HLA-DR, EGFR, CD4
- Variable positivity: CD68, S100, EMA, CD15
- Negative: CD45, CD20, CD1a, lysozyme, MPO, CD34, CD3, CD79a, CD30, HMB-45, CK
- Clusterin: sensitive and specific
- Ki-67 index: 1-25%



Treatment



- The current approach is to apply therapeutic guidelines similar to those used for soft tissue sarcomas of high grade
- Complete surgical resection is the therapy of choice
- Adjuvant radiation or chemotherapy decreases the rate of recurrence



Biologic behavior and Prognosis



- The behavior of these tumors is more akin to that of a low-grade soft tissue sarcoma than a malignant lymphoma
- Local **recurrences** rate: 36%, **metastasis**: 28%, mostly to LN and liver

Poor prognostic factors:

- intra-abdominal location
- size ≥ 6 cm
- mitotic count ≥ 5 /10 HPFs
- coagulative necrosis
- significant nuclear pleomorphism



Message?



- As spindle cell tumors within lymph nodes are rare, the diagnosis of FDCS within a node is not that difficult. However, extranodal FDCS cases are much more challenging in diagnosis because sarcomatoid carcinoma and sarcoma are by far much more common
- Clue: morphologic features



Our patient



- Patient still alive with disease
- Receiving radiotherapy...
- No surgery yet....